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Editor
HOWARD P. LEWIS, M.D.
Portland, Oregon

Associate Editor
HERBERT E. GRISWOLD, JR., M.D.
Portland, Oregon

Associate Editor
FRANKLIN J. UNDERWOOD, M.D.
Portland, Oregon

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RECOGNITION AND TREATMENT OF CONGESTIVE HEART FAILURE IN INFANCY: I. DIAGNOSIS*†

Congestive heart failure in infancy differs from the clinical picture seen in adults in its more fulminating course, more difficult diagnosis and more frequent association with acute respiratory infection. The dangerous prognosis has been stressed particularly by Dr. John Keith, of Toronto, who found approximately an 80 per cent mortality in infants under one year admitted to the Hospital for Sick Children in cardiac failure. This figure is fairly constant in any series where a large number of newborns is admitted directly from the maternity hospital, but is considerably reduced if only older infants and those referred from a distance are seen. Any newborn series will contain numerous hopelessly inoperable conditions, such as aortic atresia or cor biloculare, which will cause a high and possibly irreducible mortality. Nevertheless, there is no doubt that many older infants' lives could be saved if the diagnosis of cardiac failure were made earlier and if the management included all the recent advances in medical and surgical cardiology.

Diagnosis of the Presence of Congestive Failure

The early signs of failure are very frequently overlooked in infancy. As Keith, Rowe and Vlad have pointed out in their recent textbook, it is often the infant's grandmother who first notices and stoutly maintains, in the face of pediatric opposition, that the most cardinal of all signs is present, namely, *dyspnea at rest.* This is in part a tribute to the well-known clinical acumen of grandmothers, but in part is also a criticism of the comparative slowness and insensitivity of the maternity nurse and examining physician. There are many causes of dyspnea in the first week of life, but with each succeeding day of life the cardiac causes assume an ever more important role. The dyspnea is frequently associated with *retraction,* or indrawing of the lower ribs; suprasternal retraction is late and is more frequently due to respiratory causes, or vascular compression. A busy pediatrician or cardiologist will frequently overlook dyspnea at rest, for the simple reason that he does not see the infant at rest, but crying and fretful after numerous procedures have worn down his fragile resistance and produced the irritability so characteristic of the small sick baby. Normal respirations cannot be observed nor accurate auscultation achieved unless the infant is quiet, so that considerable time and trouble are justified in reproducing a basal state. A well-filled bottle of formula is as important as the stethoscope in such cases. Sedation with barbiturates may be used if all other measures fail.

Feeding difficulty and *poor weight gain* are characteristic of many forms of heart disease, especially the large left-to-right shunt group, and are not usually helpful in the diagnosis of failure. However, a *sudden* onset of these symptoms in an infant previously gaining well may be the first symptom of cardiac failure, and the mother should always be warned at the first examination to report such an occurrence immediately. A sudden "good" weight gain of five or six ounces a week in an infant previously almost stationary usually heralds the onset of edema. *Slowing or cessation of motor development* may be due to the onset of cardiac failure, as in one infant with pulmonary stenosis and normal aortic root who sat up at six months, started crawling at nine months, began to talk at ten months, but later became irritable, a feeding problem, then stopped crawling and was making no attempt to walk by the time he was referred for surgery in advanced cardiac failure at eighteen months.

Cardiomegaly is a cardinal sign of cardiac failure in infants, and there are only two or three conditions where it is not seen. A cardiothoracic

* From the Harriet Lane Home, Cardiac Clinic, Johns Hopkins Hospital, Baltimore, Maryland.

† Part II of this article will be concerned with the treatment of congestive heart failure in infancy and will follow in the February issue.

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ratio of 60-65 per cent or over is always present with true congestive failure, except in subdiaphragmatic anomalous pulmonary venous return, in congenital mitral valve disease and occasionally in myocarditis or paroxysmal tachycardia in the first 24 to 48 hours after their onset.

This correlation of cardiomegaly and failure may sound obvious, but despite the enormous volume of literature on congenital heart disease appearing over the past ten years, one still sees infants being treated for congestive failure with digitalis and even diuretics when they have small hearts, deep cyanosis and are clearly having cyanotic or anoxic spells due to decreased pulmonary flow. One unfortunate infant seen recently was known to have tetralogy of Fallot and was rushed to his local hospital in his first cyanotic or anoxic spell. There he was treated, despite his small heart and the fact that he lived in Maryland, where the significance of spells in "blue babies" might be thought to be appreciated, with digoxin, which was both harmless and useless, and with aminophylline which was, in his case, harmful, causing convulsions and severe gastrointestinal bleeding. Fortunately, he survived transfer and underwent a successful subclavian pulmonary anastomosis after his iatrogenic maladies had been controlled.

Although the exact physiological mechanisms underlying these anoxic spells are poorly understood, they are clearly not associated with cardiac dilatation or myocardial failure and the use of digitalis as a panacea merely delays the consideration of the need for early surgery. The only indications for diagnosing cardiac failure and using digitalis in infancy in the absence of cardiomegaly are paroxysmal tachycardia or early myocarditis. If the infant has either subdiaphragmatic pulmonary venous return or congenital mitral valve disease, his response to digitalis will be transient only. If he has tetralogy, or other malformation in the conotruncus group, there will be no response.

Gallop rhythm is frequently present and is usually protodiastolic in timing. Some experience is necessary in order to hear it and its presence should not be regarded as a *sine qua non* for the diagnosis of cardiac failure.

Hepatomegaly is almost invariably present, even when the failure is predominantly left-sided. However, since the liver of normal young infants is often palpable, since hepatic tenderness is almost never found and the position of the liver varies much more rapidly from hour to hour than it does in older subjects, depending on the degree of respiratory embarrassment, too much time is frequently spent in hospitals, where many doctors examine the same infant, in debating whether the liver is, or is not, "enlarged for the age."

When *hepatic pulsations* can be felt they invariably indicate cardiac failure. If systolic in time, they indicate tricuspid insufficiency, sometimes present in congestive failure from any cause. If presystolic in time, they indicate obstruction either at the tricuspid valve or obstruction to right ventricular outflow, and in particular that failure is probably due to pulmonic stenosis with normal aortic root. When pulsations are absent, liver size should be used as only one of many possible criteria of failure, a very distant fourth behind cardiomegaly, dyspnea and edema. A frequent error seen in teaching hospitals is the assumption by interns that the referring physician was wrong in diagnosing hepatomegaly and failure 24 to 48 hours previously, because by the time the infant arrives, fully digitalized, the liver is "little if at all enlarged." The temptation to stop digitalis and "see what happens" is too rarely resisted. If cardiomegaly and dyspnea still be present, it is wiser to assume that the hepatomegaly has responded to treatment. The significance of apparent enlargement of the liver is particularly difficult to assess in infants with large left-to-right shunts and bronchiolitis, where the diaphragm is depressed by emphysema. If any but the most minimal cardiomegaly be present it is best to assume that some degree of failure exists and treat it accordingly, at least until the acute illness has passed.

Edema is a late and ominous sign in infants. It is often missed until far advanced and then, partly because of the frequent presence of edema of the eyelids, misdiagnosed as congenital heart disease with nephrosis, a diagnosis which should never be made. The only condition in which edema is seen with a good prognosis is in the syndrome of gonadal dysgenesis and coarctation, where the extremities may show gross lymphedema but no other signs of failure are present.

Pulmonary edema is rare in florid form, but carries the same urgent indication for treatment as in adults. The wheezing element is more pronounced than the fine crepitations heard in adults, but the irritating cough, the restlessness and the ashen color are similar. The diagnosis from bronchiolitis may again prove difficult and indeed the two conditions may coexist. A concept useful to many of us is that subclinical pulmonary edema is constantly present in most infants with large left-to-right shunts or with transposition of the great vessels, and this accounts in part for their frequent respiratory infection. It was shown experimentally by Dr. Barry Wood, and others, studying the susceptibility of the lung to secondary infection after influenza virus, that pulmonary edema fluid is not only an excellent culture medium, but its presence impairs the normal phagocytic processes in the pulmonary alveoli.

Any acute respiratory infection is likely to precipitate these infants into acute cardiac failure. Rarely, in congenital mitral stenosis, acute transient attacks of pulmonary edema may be produced by feeding and relieved by rest and the

Table I

Causes of Congestive Cardiac Failure in Infancy

I. Left-to-Right Shunts

A. Intracardiac

1. Atrial level
   - Atrial septal defect*
   - Anomalous pulmonary venous return (supradiaphragmatic)*
2. Ventricular or atrioventricular level
   - Atrioventricularis communis, ostium primum†
   - Ventricular septal defect†
   - Single ventricle without pulmonic stenosis

B. Extracardiac

1. Truncus arteriosus
2. Aortic pulmonary septal defect*
3. Patent ductus arteriosus*

II. Obstruction to Ventricular Outflow

A. Left ventricle

1. Aortic stenosis*
2. Coarctation of the aorta†

B. Right ventricle

1. Pulmonic stenosis with normal aortic root*
2. Primary pulmonary hypertension
3. Ebstein's malformation of the tricuspid valve

III. Obstruction to Pulmonary Venous Return

A. Cor triatriatum*
B. Congenital pulmonary venous stenosis
C. Anomalous pulmonary venous return (subdiaphragmatic)
D. Congenital mitral insufficiency
E. Congenital mitral stenosis*

IV. Transposition of the Great Vessels

A. Complete†
B. Partial
   Example: Taussig-Bing malformation

V. "Myocardial" Group

A. Anoxic

1. Anomalous origin of left coronary artery from pulmonary artery**
2. Coronary calcinosis
3. Endocardial fibroelastosis**

B. Infiltrative

1. Glycogen storage (Pompe's disease)
2. Tumors

C. Infective

1. Myocarditis
2. Pericarditis

D. Dysrhythmic, etc.

1. Paroxysmal tachycardia
2. Congenital heart block
3. Thyrotoxicosis, natural or drug-induced

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* Operable; surgery nearly always indicated.
† Operation available, but needs much debate in individual case.
** Operation available, but hazardous or unsatisfactory.

upright position. Pulmonary edema may also occur immediately postoperatively following relief of severe pulmonic stenosis.

Venous engorgement is left to the last, not because it does not occur, but because these infants are frequently so thin that their veins appear prominent even in the absence of failure, and large engorged veins are often seen in the head and neck in such conditions as tetralogy, where congestive failure does not occur.

Thus, although the signs and symptoms of congestive cardiac failure in infants do not differ essentially from those in older children and adults, the relative significance of each, and the ease with which they can be detected, are often different and thus can give rise to frequent difficulty.

Diagnosis of the Cause of Cardiac Failure

Some bizarre malformations, such as aortic atresia, cor biloculare, and others, cause death in the first week of life and no therapy, medical or surgical, is at present of any avail. It is bet-

Table II

Clinical Diagnosis of Congestive Failure in Infancy

Diagnostic Groups	I Left-to-Right Shunts	II Obstruction to Ventricular Outflow	III Obstruction to Pulmonary Venous Return	IV Transposition of the Great Vessels	V "Myocardial" Group
CYANOSIS	Mild, often only at birth or at onset of failure	Mild to moderate in pulmonic stenosis only	Variable	Deep and persistent	Absent except in late failure
SYSTOLIC MURMUR	Present	Basal, stenotic in type	Presystolic or mid-diastolic in mitral stenosis	Inconstant	Absent or insignificant
PULMONIC SECOND SOUND	Increased	Decreased in pulmonic stenosis—A_2 decreased in aortic stenosis	Increased	Increased	Normal
PULMONARY VASCULARITY	Increased	Decreased in pulmonic stenosis—normal in others	Fine mottling or edema	Increased	Normal
ELECTRO-CARDIOGRAM	RVH only in atrial shunts—LVH, or combined in others	RVH and S-T depression in right ventricular obstruction—LVH in aortic stenosis—variable in others	RVH	RVH, or combined	T waves flat or inverted in left precordial leads—LVH in endocardial fibroelastosis

ter, at this time, to concentrate such diagnostic acumen as we possess on those conditions which are both better defined and offer some hope of cure. I do not by any means imply that the investigation of these infants is fruitless or that they should be denied medical treatment, but if one excepts them from the present discussion, then the causes of congestive failure in infancy can be said to fall into five well-defined groups and a logical approach to differential diagnosis can be made.

Although an exact anatomical diagnosis cannot always be made without special studies, in the large majority of cases it is possible to decide, on the basis of history, physical examination, x-ray, fluoroscopy and electrocardiogram, which of the major groups above is the cause of failure in a particular infant (Table I).

I. *Diagnosis of a Left-to-Right Shunt:* The *history* is typically one of dyspnea, feeding difficulty and poor weight gain from the beginning, becoming more marked with the onset of failure. Cyanosis may be previously minimal or absent, but appear later with the onset of failure. Serious errors are often made and the relatively hopeless diagnosis of transposition reached because the infant is cyanotic when first seen. With rare exceptions, the cyanosis of transposition is deep, constant, present from birth and admitted by all to be present. "Controversial" cyanosis, seen by some and not by others and inconstant from day to day, is by no means incompatible with a large left-to-right shunt and is particularly common in anomalous pulmonary venous return. *Examination* reveals a systolic murmur in almost all cases and an accentuated pulmonary second sound. The harsh holosystolic murmur and thrill of a ventricular shunt, the diffuse blowing murmur of an atrial shunt and the continuous murmur of a ductus may all be recognized, even in a young infant. The bounding pulses and wide pulse pressure of a patent ductus contrast as vividly as in the adult with the small pulse and turbulent precordium characteristic of atrial shunts. *X-ray and fluoroscopy* reveal increased pulmonary vascularity and hilar pulsations. In far advanced heart failure from any cause there may be passive hilar congestion due to pulmonary edema, and the status of the pulmonary arteries may be difficult to determine. X-ray and fluoroscopy may then need to be repeated a day or two after the failure has improved. The most frequent difficulties, however, are often technical, where full inspiration, a large thymus and poor technique together produce the so-called "fog-at-sea" type of film. Since an accurate assessment of pulmonary vascularity is essential, no effort should be made to read such films and the infant should be transferred as early as possible to a center with adequate facilities. It is rarely that an experienced reader of a good film will make an error as to the presence or absence of a shunt, but if there is any doubt at all, catheterization should be performed. *Electrocardiography* is useful, indeed vital, in localizing the shunt. Atrial shunts are characterized by right axis deviation, right atrial and right ventricular hypertrophy and complete absence of left atrial and left ventricular hypertrophy. Ventricular and extracardiac shunts are characterized by variable axis deviation, combined ventricular hypertrophy and frequently, as Ziegler has demonstrated, by left atrial hypertrophy.

II. *Diagnosis of Obstruction to Ventricular Outflow:* These patients usually have normal growth and development until the onset of cardiac failure. Attacks of anginal pain may rarely be present in aortic stenosis, and are similar to the attacks to be described later in anomalous origin of the left coronary artery from the pulmonary artery. *Examination* in aortic or pulmonic stenosis reveals a basal systolic murmur of varying intensity, stenotic in character, that builds up to a crescendo at the second heart sound and is frequently accompanied by a thrill. The corresponding second sound is diminished or absent. In pulmonic stenosis, presystolic hepatic pulsations and giant "a" waves in the jugular pulse are often present. *X-ray and fluoroscopy* in pulmonic stenosis reveal gross right atrial and right ventricular enlargement and normal or diminished pulmonary vascularity. In aortic stenosis, the enlargement is entirely left ventricular, and the pulmonary vascularity normal. The *electrocardiogram* shows right axis deviation and gross right atrial and right ventricular hypertrophy with S-T depression in pulmonic stenosis. In aortic stenosis the axis is usually normal and the precordial leads show left ventricular hypertrophy with T-wave inversion in Leads 1, V_5 and V_6.

The differential diagnosis of pulmonic stenosis from the other causes of right ventricular outflow obstruction may be difficult on clinical grounds alone; one's management of the case must include the consideration that pulmonic stenosis is the most frequently encountered condition and the only operable one.

Coarctation of the aorta is recognized by a difference in pulse volume and blood pressure between the upper and lower extremities. It is essential to palpate brachial and femoral pulses simultaneously for two reasons: first, in profound failure all the pulses may be weak and if the femorals alone are palpated, coarctation may be diagnosed when not present; and second, if one feels sufficiently long with sensitive fingers, *some* femoral pulsation can always be felt. The diagnosis is based on the discrepancy between the bounding, often readily visible, pulsations of the brachial artery compared with the weak or absent femorals. The difference in blood pressure between upper and lower extremities depends on the type of coarctation, being less in the

infantile or preductal than in the adult or postductal type, and on the degree of failure, being less pronounced when failure is far advanced. The systolic pressure in the arms by the flush technique is usually at least 20 mm. above that in the legs, and may be 100 mm., or more. Cardiac murmurs are insignificant and variable unless the coarctation be accompanied by some other malformation. *X-ray and fluoroscopy* reveal normal vascularity and enlargement of the left ventricle, the other chambers being normal. Rib notching and the "3" sign are not seen in infancy. Crafoord's sign is very rare, although if present is helpful in localizing the coarctation. The *electrocardiogram* is inconstant and nonspecific, since it may show left or right ventricular hypertrophy, depending apparently on the age at which symptoms develop.

Normally, infants with coarctation grow well until the onset of failure. Exceptions are gonadal dysgenesis and one case, vividly remembered, of abdominal coarctation immediately above the renal arteries, where presumably impaired renal function caused the remarkably stunted growth.

III. *Diagnosis of Obstruction to Pulmonary Venous Return:* The rare malformation, anomalous return of the pulmonary vein below the diaphragm into either the portal vein or the hepatic vein, has been classified here because the whole course of the infant's brief life appears to be one associated with obstruction of the venous return at its entrance into the hepatic or portal vein rather than the characteristic picture of left-to-right shunt. These infants show intermittent cyanosis, very little, if any, murmur, increasing right ventricular hypertrophy on the electrocardiogram and, as mentioned earlier, a heart which is very little, if at all, enlarged in the presence of gross hepatomegaly, dyspnea and other signs suggestive of cardiac failure. The lung fields are said to show a rather characteristic mottled type of appearance. The clinical syndrome has recently been well elucidated by Johnson, and his associates, in Montreal. Cor triatriatum and congenital mitral stenosis may both behave very similarly in that the infant has recurrent attacks of pulmonary edema, reveals gross right ventricular hypertrophy on the electrocardiogram and may or may not show a good presystolic murmur suggesting mitral valve obstruction. In one case seen here the murmurs were indeed classical for mitral stenosis, even to the presence of a well-marked opening snap, but this has by no means always been the case in the other infants recorded in the literature. One should probably think of this diagnosis in the presence of recurrent attacks of pulmonary edema, or in any case with no suggestion of a left-to-right shunt and signs of pulmonary hypertension. Dr. Ferencz recently analyzed the literature and pointed out how frequently congenital mitral stenosis was associated with other cardiac anomalies. When, however, it is present as an isolated phenomenon, a gratifying result can be obtained by mitral valvotomy, although this is a technically difficult procedure. Congenital mitral insufficiency is an even rarer cause. It is a well recognized condition in infancy, causing, as in adults, enlargement of the left atrium and a blowing systolic murmur maximal at the apex and conducted out to the axilla.

IV. *Diagnosis of Transposition:* Deep and persistent cyanosis dating from birth, gross failure to thrive, the early onset of cardiac failure and the characteristic "egg on its side" shape of the heart, make this one of the easiest, most frequent and most tragic of diagnoses. Murmurs are inconstant, depending on the associated defects in the heart. The pulmonary vascularity is greatly increased and the electrocardiogram shows right or combined ventricular hypertrophy.

V. *Diagnosis of "Myocardial Failure":* The most rewarding of this group is *paroxysmal tachycardia,* diagnosed by a cardiac rate of 180 or more, and typical superimposition of P and T waves in the electrocardiogram. Nadas has commented on the frequent association of supraventricular tachycardia with intraventricular block in children, giving a bizarre appearance that may be erroneously diagnosed as ventricular tachycardia. *Myocarditis* may accompany an obvious respiratory infection, or may appear *de novo.* The infant is perfectly healthy until the onset of failure over the course of one or two days. The correct diagnosis is suggested by the sudden onset, poor quality heart sounds, gallop rhythm and electrocardiogram showing flattening or inversion of the left precordial T waves with little or no evidence of left ventricular hypertrophy. The absence of left ventricular and left atrial enlargement helps in distinguishing this from *endocardial fibroelastosis.* The latter is a common condition but is being diagnosed too often. It is a "safe" diagnosis, in that it is difficult to disprove except by autopsy. The comparatively recent recognition of a similar condition in adults has led many to attach this diagnosis to a medley of cardiac failures in infancy. Although the diagnosis may be suspected clinically from the striking left atrial and left ventricular enlargement without obvious cause, it should rarely be firmly made without ancillary diagnostic aids.

Anomalous origin of the left coronary artery from the pulmonary artery is differentiated from the above condition partly by the age of onset of symptoms which appear almost invariably at about three or four months of age and consist of dyspnea so gross as often to lead to a diagnosis of pneumonia and episodes of angina. The anginal episodes are accompanied by screaming,

drawing up of the legs on to the abdomen, profuse sweating, pallor and complete collapse. The electrocardiogram typically shows an anterior infarction or Q_1T_1 type of pattern. When, as occasionally happens, this pattern is absent, the diagnostic difficulties are greatly increased.

All of this latter group are characterized by comparatively normal development until the onset of symptoms, by an absence or paucity of murmurs, combined with left ventricular hypertrophy of varying degrees and the absence of deep cyanosis or any sign of shunt.

CATHERINE A. NEILL, M.D., M.R.C.P.
First Assistant to Helen B. Taussig, M.D.
Harriet Lane Home, Cardiac Clinic
Johns Hopkins Hospital, and
Assistant Professor of Pediatrics
Johns Hopkins University
Baltimore, Maryland

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